

Progressive Pan-Corporal Hemi- Hypertrophy: A Case Presentation



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Abstract

Hemi-hypertrophy is a rare phenomenon affects most organs mostly musculoskeletal system. A teenager lady was admitted to have operations on left sided hypertrophy in face and trunk and lower limb macrodactyly. Involved area had extra adipose and connective tissues with normal bony structures. She had not any sign of visceral malignancies. Overall, she did not contained all criteria of hypertrophy syndromes or Isolated Hypertrophy.

Introduction

The hemi-hypertrophy is one of unusual abnormalities can be congenital which affects multi systems. Musculoskeletal involvement characterized as variable growth in different parts of body. It can be assumed as a part of syndromes like Beckwith-Wiedemann syndrome, Proteus syndrome, Klippel Trenaunay Weber syndrome or Hemi-hyperplasia- Multiple lipomatosis or can be happened as idiopathic event [1,2]. Tumors like Wilms and hepatoblastoma are among common tumors found in isolated hemi-hypertrophies [3]. Management of hemi- hypertrophy is usually conservative and according to encountered conditions can plan. We want to present a case of hemi-hypertrophy who had multiple pictures of such phenomenon.

Case Presentation

An 18 year's old lady consulted for her appearance deformities. The problem was noticed from delivery, but progressively asymmetry changed her status. Her IQ went beyond average level, as well she had not any history of seizure or behavior problems. She was concern about left side facial hypertrophy which caused deviated upper lip and mallar area, as bizarre jowling face. However, facial hard tissue component seemed structurally and functionally normal (Figure 1). On the other hand, hypertrophic left side thoracolumbar subcutaneous tissues changed the posture. So, she suffered kyphoscoliosis with limitations in her carriers (Figure 2). In the lower limb, macrodactyly of some toes was significant bilaterally (Figure 3). In the reports of laboratory tests, there was not any abnormal values. Previous imaging studies demonstrated no sign of malignancy or visceral masses. The patient underwent

two steps of debulking in face and trunk to decrease dysregulated adipose tissue volume.

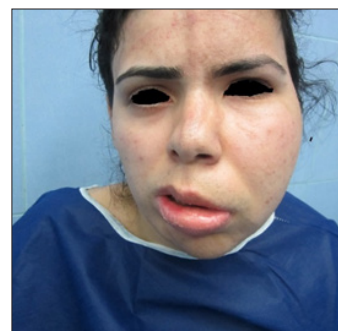


Figure 1: left sided facial hemihypertrophy without malocclusion.



Figure 2: left sided hypertrophy of subcutaneous tissues of Trunk with Café au lait patches.



Figure 3: lower limb macrodactyly-especially in middle toes.

Discussion

Overgrowth of different parts of body is the main-stem in hypertrophy or hyperplasia events. Syndromic case are generally associated with genomic mutations affect structural proteins regulation [1]. Klippel Trenaunay Weber syndrome, Proteus syndrome, Beckwith-Wiedmann syndrome are among common syndromic forms of such hypertrophies [4]. Moreover, isolated hypertrophies (IH) are potential sources for tumors and knowledge about their diagnostic criteria, can be useful to manage if not life-saving. So, Clericuzio introduced some criteria to diagnose and illustrated association between IH and tumors, though we know these are not exclusive [5]. In some isolated forms there are

associations with genomic informations like chromosome 11p15, as shown by Shuman et al. [6]. Beyond this information, our case did not contained all criteria of each of syndromic forms nor presented isolated hyper trophy. She had complained deformed face and trunk and microscopic findings of surgery's products following facial and trunk debulking were compatible with adipose and connective tissue. Illustration of clinical manifestations of bizarre postures, help new colleagues to determine their unknown patients and choose better decisions.

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